Intracranial spread of intramedullary spinal cord ependymoma: Report of two cases

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Spinal dissemination of intracranial ependymomas through cerebrospinal pathways is not rare and reported to occur in approximately 10% of the cases. However, the reverse is an uncommon phenomenon. We report two children who were initially treated for spinal ependymoma but presented later with multiple intracranial metastases. [Turk J Cancer 2000;30(3):119-125]

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The spread of ependymoma from brain to spinal canal occurs in approximately 10% of all cases (1). Dissemination of the neoplastic cells within the ventricular cavity is thought to be facilitated by the active circulation of the cerebrospinal fluid (CSF) throughout the neuraxis, coupled with the effect of gravity (2). However, the reverse is an uncommon phenomenon (3). We report here two young patients with intracranially disseminated spinal ependymomas.

Case 1

A 12-year-old boy was admitted with the complaints of weakness and pain in both legs in September 1987. His neurological examination revealed motor and sensory paresis below T10 level. Computerized tomography (CT) examination demonstrated an infiltrative intramedullary mass without syrinx and a widening of the spinal canal between T11 and L3 segments. The patient underwent a gross total excision of intramedullary lesion through L1-4 laminectomy. Pathological examination was reported as grade–II ependymoma. Motor function slightly improved following surgery. A total of 49.50 Gy radiotherapy in 33 divided doses (1.5 Gy/day) was delivered to T10–L5 area through single posterior field by Cobalt–60 teletherapy unit. Five years later, the patient developed progressive paraparesis and CT and CT/myelography showed an intramedullary tumoral mass at T2–T4 levels. A laminectomy was performed and gross total removal was achieved. Histopathological diagnosis was once more ependymoma grade–II.
The patient was started on a chemotherapy regimen of vincristine, procarbazine, CCNU and dexamethasone for 11 consecutive courses.

In July 1996, he was referred again with progressive neurological deterioration for radiotherapy of multiple tumoral lesions of spinal segments T5-10 (Figures 1,2). Radiation dose of 30 Gy was given in 10 fractions to T4-10 spinal segments that made no recovery in neurological deficit. In December 1997, the patient was admitted to our department with the complaint of additional progressive neurological symptoms of nausea, vomiting and partial hearing loss. Magnetic resonance imaging (MRI) examination showed multiple new intracranial and spinal seeding metastases (Figure 3). The patient received a new course of radiation therapy of 52.2 Gy to cranial lesions and 30.6 Gy in conventional fractions to spinal segments not irradiated before. The patient was discharged after the radiotherapy sessions and was reported to be dead one month later.

Fig 1. Post-gadolinium, T-1 weighed sagittal MRI of the spinal cord of case 1, showing recurrent tumor at the previously operated T9–11 segments and kyphotic deformity
Fig 2. Post-gadolinium, T-1 weighed sagittal MRI of the spinal cord of case 1, showing tumoral lesions wide spread at spinal segments T5-T10, multiple subarachnoid seeding metastases at T2-T3, C1 and C3

Fig 3. Post-gadolinium, T-1 weighed sagittal MRI of the cranium of case 1 reveals multiple infratentorial enhancing lesions with a large suprasellar mass consistent with seeding
Case 2

In January 1995, a 2.5 years old girl was admitted to Hacettepe University Hospitals with a two–months history of weakness of both legs and arms that was more prominent in the right arm. The family also described restlessness and anorexia for the last month. There was no sphincteric disturbance. Neurological examination revealed spastic quadriplegia more prominent on the right side. MRI examination revealed an intramedullary tumoral mass at C3-7 level. The patient had a gross total excision of the tumoral mass through posterior cervical route. Histopathological examination revealed grade–II ependymoma. The child’s quadriplegia markedly improved with only mild weakness of the right extremities. A total dose of 42 Gy over 36 days was administered to tumor bed using 15 MeV electron beam. The patient did well during radiotherapy. After radiotherapy, six cycles of chemotherapy schedule including vincristine, CCNU and procarbazine was administered to the patient. MRI during follow–up period, confirmed no residual tumor (Figure 4).

Fig 4. Post-gadolinium, T-1 weighed sagittal MRI taken one year after initial diagnosis of case 2 shows no residual disease at the primary tumor site
In September 1996, the patient developed symptoms of diabetes insipidus. MRI investigation revealed a suprasellar tumoral mass compressing the anterior recess on the third ventricle and hypothalamus which was not present at the corresponding MRI section previously. Nasal desmopressin (Minirin®) treatment was started. Three months later, repeated MRI examination demonstrated an increase in the size of intracranial mass and multiple new seeding metastasis in preoptic and ambient cisterns (Figure 5). There was no recurrent lesion in the cervical spinal region. The patient underwent an operation for subtotal excision of the suprasellar tumor. The histopathological diagnosis was reported as grade–II ependymoma. In the postoperative period, she received 55 Gy radiotherapy to cranial lesions by 6 MV X-rays. The patient died of progressive disease 42 months after the initial diagnosis.

Fig 5. Post-gadolinium, T-1 weighed sagittal MRI of case 2, two years after initial diagnosis demonstrates a suprasellar homogenously enhancing mass obstructing the anterior recess of the third ventricle. There are multiple enhancing areas at pontine surface and 4th ventricular ependymoma consistent with subarachnoidal seeding. There is no recurrent lesion at the primary tumor site.

Discussion

Spinal cord ependymomas comprise less than 2% of all central nervous system neoplasms, 15% of spinal cord tumors and up to 60% spinal cord
gliomas (4). Surgery is the treatment choice for intramedullary ependymomas. Total excision of the tumor, especially in low-grade ependymomas, leads to excellent results (5). However, incompletely resected high-grade spinal ependymomas have a dismal prognosis. Postoperative radiation therapy has improved the outlook for these patients significantly. It has played an important role in local control and prolonging life by increasing the 5-year survival estimates three-fold (6).

Spread of intracranial tumors to the spinal canal, especially high grade ones, is a well-known entity. In spinal ependymoma series, most of the treatment failures have been reported to be local, whereas distant metastases and dissemination through cerebrospinal fluid (CSF) have been rarely documented (3,6,7). Rezai et al. (1) reported that 11.4% of all intracranial ependymoma cases were disseminated in the central nervous system. In a review of 14 spinal ependymoma series from the literature, Whitaker et al. (5) calculated a 5.8% incidence of cranial relapse in 259 patients. Waldoron et al. (3) reported 2 patients out of 59 who relapsed within the brain.

We consider our two patients as intracranial dissemination of spinal ependymoma. In the second case, the MRI revealed a tumoral mass in the ventricle which was previously normal. The time course for the progression of symptoms in the first case makes it unlikely that the primary lesion was intracranial, or that the lesions were multifocal initially.

Analysis of 140 patients by Rezai et al. (1) revealed that relatively younger patients were at risk for dissemination of the primary tumor during their clinical course. In previous reports, the patients with intracranial tumors showing spinal cord dissemination were generally young with an age range from 5 to 48 years (median 16.8 years). To our knowledge, case two is the youngest patient reported to date. Although the multiple operations on intramedullary spinal cord tumors has been proposed as a risk factor for spinal and cranial dissemination, it is difficult to assess the association statistically (2). In case 2, total excision of initial tumor and one single operation before the dissemination is not consistent with this possible risk factor, whereas multiple surgical interventions with incomplete resections in the first case is.

There are conflicting data in the literature regarding the correlation of ependymoma histology with clinical prognosis. It has been shown that high grade ependymomas disseminate more commonly throughout the CNS. Whitaker et al. (5) reported 3 cases of intracranially disseminated ependymoma, two of them received craniospinal irradiation due to high-grade tumor. Shaw et al. (7) reported one adult patient with grade 3 disease who disseminated intracranially, out of 22 patients with spinal ependymoma. In other series, Waldoron et al. (3) reported two (2/59) intracranial failures, both with anaplastic tumors. Our cases, being grade-II are not consistent with previous reports.

In conclusion, although rare, it should be noted and kept in mind that spinal ependymomas may disseminate into the cranium especially in high risk cases. A complete radiographic evaluation of the entire neuraxis should be performed in the initial work-up of these cases and the treatment should be planned accordingly.
References